



Review

The psychosocial and psychiatric side of cystic fibrosis in adolescents and adults

P.E. Pfeffer, J.M. Pfeffer*, M.E. Hodson

Department of Cystic Fibrosis, Royal Brompton and Harefield NHS Trust, Sydney Street, London SW3 6NP, UK

Abstract

Increasing numbers of cystic fibrosis (CF) patients are surviving into adulthood. An understanding of the psychiatric and psychosocial aspects of CF in adults and adolescents is therefore more important than ever. There is a large body of evidence indicating that the psychological and psychosocial functioning of people with CF is similar to that of well people, until the disease becomes severe. However, there is also evidence that patients do suffer an increased likelihood of psychiatric problems, such as depression, and of scoring poorly on physical functioning measures of quality of life. Studies have found conflicting evidence as to any association between degree of respiratory impairment and psychological functioning. Coping styles seem to have a large effect upon the quality of life of CF patients. People with cystic fibrosis can have problems with sexuality, platonic relationships and independence. Families of patients also suffer problems, which can affect the patients themselves. Non-compliance is a complicated problem with many patients. New treatments for people with CF are emerging, such as lobe transplants from live donors and gene therapy, with possible new psychosocial problems resulting. Furthermore, older studies are becoming increasingly inapplicable as treatment and prognosis changes. Therefore, more research is needed in this field.

© 2003 European Cystic Fibrosis Society. Published by Elsevier Science B.V. All rights reserved.

Keywords: Cystic Fibrosis; Psychology; Psychiatry; Compliance

1. Introduction

The prognosis of cystic fibrosis has changed dramatically over the last 10 years or so with increasing numbers of patients surviving into adulthood. It is important not to overlook the psychosocial aspects of this group of patients to ensure the quality of their increased life is good. In order to do so, it is necessary first of all to try and assess what level of psychosocial problems are present and, as a first step, we have reviewed the literature in this area. With the changing prognosis many older psychosocial studies may no longer be appropriate and we have therefore concentrated on more recent papers (from 1990 onwards).

2. Quality of life in people with Cystic Fibrosis

Although a corner stone of modern medicine, the term ‘quality of life’ lacks a clear definition, but is often taken as a measure of the impact of a disease on

psychological, psychosocial and physical functioning. Varying emphasis on these different aspects of functioning effects whether cystic fibrosis is found to lead to a significantly worse quality of life (QOL) or not.

A number of studies have concentrated on the psychosocial and psychiatric impact of cystic fibrosis. Anderson et al. [1] found only some subclinically elevated levels of psychopathology (hypochondriasis and hysteria on Minnesota Multiphasic Personality Inventory –2) in cystic fibrosis adults, though the prevalence of clinical depression and anxiety was similar to the general population. However, the different analytical instruments used in this study found conflicting evidence of which factors may be associated with more serious psychopathology. For example, male gender was found to predict significantly higher levels of depression using one psychological test (Beck Depression Index) but not another (Minnesota Multiphasic Personality Inventory –2). These conflicting results using different psychiatric instruments question whether these tests are suitable for accurately discerning the levels of psychopathology in cystic fibrosis or may indicate that the levels of psycho-

*Corresponding author..

pathology are of borderline significance. The two instruments do have slightly different focuses—the former focusing on mood in the preceding 2 weeks and the latter more on personality traits.

Pearson et al. [2] tested adults with cystic fibrosis and found a prevalence of anxiety of 22.2% and depression of 42.4%. They found that the different indices of psychiatric behaviour and global emotional difficulties were intercorrelated. Blair et al. [3] found the psychological functioning of young adults with cystic fibrosis to be in many ways similar to that of healthy young people.

Shepherd et al. [4] and de Jong et al. [5] found that the psychosocial functioning of CF patients did not differ significantly from that of healthy control groups. However, de Jong et al. [5] did find the cystic fibrosis patients to have a significantly worse quality of life reflecting their significantly impaired scores for physical functioning. Shepherd et al. [4] found some differences between CF patients and controls, particularly in the area of undertaking life-style changes, and similarly, Blair et al. [3] found that CF patients were less likely to be in employment than subjects in the control group. Congleton et al. [6] found that although patients with cystic fibrosis were more likely to report problems in areas of daily living than a normal population, their QOL was generally similar to that of patients with minor non-acute health conditions, not being nearly as impaired as might be expected. Recently, Britto et al. [7] found young adults with CF to have lower scores on several measures of physical functioning and for general health perception, but to have similar scores for most psychosocial measures.

Therefore, on the whole, the evidence supports young adult CF patients generally having a psychological functioning similar to healthy controls. However, CF does have a negative impact on physical functioning aspects of quality of life. Nevertheless, a large proportion of adults with CF are living full and productive lives [8].

3. Relation between clinical health and quality of life

An important question is whether there is a relation between clinical health measures and quality of life. This can be subdivided into whether clinical health measures are predictive of QOL, whether acute changes in clinical health are associated with changes in the QOL and whether the QOL of patients changes over the long-term as their health deteriorates.

There is conflicting evidence as to whether clinical health measures are predictive of quality of life, though there is little study of patients who are severely ill and rapidly deteriorating. Anderson et al. [1] found no significant differences in psychological functioning between subgroups of patients based on lung function

or body weight, though a MANOVA statistical procedure found better health status to predict lower scores of psychological distress. Pearson et al. [2] found no significant correlation between duration or severity of illness and psychiatric symptomatology (anxiety, depression, eating disorders, or global emotional disturbance). Britto et al. [7] found that there was no significant association between physical/psychosocial scores and lung function or exercise tests. The relevance of this finding to adults is questionable as the young adult subgroup was combined with data from younger patients for the analysis of QOL associations and predictors. However, de Jong et al. [5] found that although QOL scores were not significantly related to pulmonary function tests, maximal exercise capacity was negatively correlated with overall and physical QOL impairment. Furthermore, MRC dyspnoea (Medical Research Council graded scale of limitation in function due to dyspnoea) correlated highly with overall, physical and psychosocial quality of life scores. Congleton et al. [6] found there were correlations between the scores in many of the dimensions (for example energy, sleep and physical mobility) which assess perceived distress/disability and clinical variables such as FEV₁% predicted, shortness of breath and time in hospital, but not body mass index. Orenstein et al. [9] found that QOL scores, measured using the Quality of Well-Being scale, were significantly related to pulmonary function tests and exercise performance, which is not surprising as the QWB scale does emphasise physical functioning in judging QOL. The QWB scale assesses recent health-related limitation of mobility, physical activity and social activity, and recent problematic symptoms.

There is evidence that acute changes in health do have an effect on quality of life. Britto et al. [7] found that an increasing number of pulmonary exacerbations in the previous 6 months and a decreasing number of days since the last exacerbation both had a detrimental effect on a patient's QOL. In a second study, Orenstein et al. [10] also found improvements in overall QOL (QWB scale) correlated with improvements of most pulmonary function tests during treatment of an acute pulmonary exacerbation in CF adolescents. Our experience at the Brompton is that mood deteriorates with serious decline in physical health after a long period of stability.

There is little research into the whether long-term changes in health effect quality of life, either in terms of a cohort of patients or the individual. A follow-up study to Congleton et al. found poor correlation between changes in physiological parameters and changes in Nottingham Health Profile scores over a small number of years (1–3) [11]. However, these patients were in a relatively stable phase of the disease. There was a significant correlation between changes in the energy and physical mobility dimensions and change in a 'CF-

specific' symptom-based total score over the years studied.

One possible explanation of why there appears to be little consistent correlation between somatic health measures and mental health measures is the discrepancy in some patients between their subjective health perception and their health as measured by clinical scales. Staab et al. [12] found that although clinical medical scores, hours spent daily on treatment and subjective health perception all were important explanations of variance in QOL in a group of 89 adult and adolescent patients, subjective health perception was the strongest predictor. Subjective health perception often differs from clinical health as measured by physicians [13,14].

The long-term adaptation and coping strategies employed by cystic fibrosis patients may well also partly explain the lack of association between quality of life and disease severity—as the disease severity slowly changes over the long-term the patient adapts, so their QOL does not decline until the disease becomes so severe they cannot adapt successfully. However, large changes in the physical health over short time-periods may lead to changes in the patient's QOL before the patients adapts. Change of disease severity, and rate of change, rather than absolute disease severity, may well be more likely to correlate to level of psychological/psychosocial functioning. Therefore, longitudinal studies of the psychological/psychosocial health of patients are needed. Another explanation of the apparent lack of correlation between psychological/psychosocial health and disease severity may be that the variance in social support may hide any relationship to disease severity. Anderson et al. [1] did find that better psychosocial support was a strong indicator of better psychological functioning.

All these studies have used instruments designed for general medical problems. One of the problems in comparing studies is that they have all used different analytical instruments. Several CF-specific QOL instruments are being designed, for example Gee et al. [15] which may be more sensitive in detecting changes in the QOL of CF patients. However, standardising disease-specific instruments may be difficult and there is the danger in such instrument being overly-sensitive and not weighting appropriately the issues which actually affect the patients' quality of life the most.

Finally, the variance in QOL amongst cystic fibrosis patients is likely to be affected by many variables and, therefore, finding a strong correlation between QOL and any single variable, without accounting for other variables, would seem unlikely.

4. The effect of gender

Although older papers suggested that women were more likely to suffer severe emotional disturbance than

men, the limited number of more recent papers do not necessarily support this. In fact, Anderson et al. [1] found that male gender was, if anything, a predictor of psychiatric emotional disturbance. Men with CF may be especially liable to suffer psychological distress in being unable to fulfil the traditional male role of being the provider [8]. The differences in the emotional problems of the two sexes could partly be a result of how Anglo-Saxon culture effects them.

Congleton et al. [6] reported that some of the dimensions in which men and women suffered distress/disability were shared (e.g. pain) and others different (e.g. men in the dimension of energy, while for women sleep). Similarly, the areas of daily living that the two genders reported problems in were partly shared and partly different. For men, there was an age-related trend in several dimensions with perceived quality of life in older men increasingly worse than scores in a healthy population of the same age. There were no statistically significant age-related trends in women.

Coping patterns also differ and this may effect long-term health [16]. Female adolescents rely heavily on denial. Males rely less heavily on denial but have more behavioural problems. Males also appear to integrate better having a physical disorder into their self-concept. Female patients remain longer in hospital than male patients even though their physical problems are no more severe [16].

Willis et al. [17] have suggested that the interaction between the social identities of the two genders and management of cystic fibrosis may be important in explaining a body of evidence that there is a higher morbidity and mortality for cystic fibrosis in females. The feminine identity may invoke a more passive approach to health and less investment in the future. The masculine identity may encourage a more active approach to health together with less cognitive preoccupation with the negative aspects of the disease upon their lives. Furthermore, the social pressure of women being slim may encourage poor nutrition in female patients. Beneficial exercise may be promoted in male patients by the social norm of a preoccupation with sport and fitness in men, though not so in women. However, data from our unit looking at morbidity and mortality over the last 40 years show no gender difference.

5. The effect of age

When considering the psychosocial functioning of people of different ages it is difficult to distinguish between the effect of ageing itself and the effects of the changing prognosis, treatments and attitudes of different patient cohorts, as outlook has improved.

Adolescent patients with cystic fibrosis may well suffer emotional disturbance both as a result of the

disease and the general emotional turbulence of adolescence. One patient reflecting on his own experience [18] commented that adolescents may be annoyed by constant questioning of how they feel (reminding them of their illness), and the lengthy daily treatment. There was a danger of developing a defence mechanism of social isolation to avoid distress from comparing themselves to healthy peers. Adolescents may also be annoyed by referral to paediatric centres [19].

Older literature remarks that small stature and delayed puberty may cause depression, poor body-image, strained interpersonal relationships, increased awareness of death and denial of sexuality [20,21]. There is little modern literature on whether adolescent or adult patients do now suffer psychological distress resulting from small stature and delayed puberty—better treatment during childhood and adolescence may well mean that these are now less severe problems. The possibility of short stature and delayed puberty should be explained to teenagers with CF before they get worried about such problems [22].

Pearson et al. [2] found eating disorders to be more common in children and young adolescents with CF than in adults, although this may merely reflect the natural history of eating disorders. A recent study found no increase in eating disorders in CF adolescents when compared to a control group [23].

The transfer of a patient from a paediatric CF centre to an adult centre is a time of great importance in the treatment of the patient—it may generate anxiety in patients and some may wish not to move from the centre they are familiar with. Non-compliance may occur [24]. Transfer is best done with joint sessions. Treatment of young adults on paediatric wards sometimes engenders the childish reaction of rebelliousness owing to the less accommodating nature of paediatric wards [25].

As patients get older and their disease more serious, studies have found that their self-esteem declines, they suffer from more discomfort with their social situation, increased frustration and their compliance decreases. Pearson et al. [2] found older patients to have a higher prevalence of anxiety and depression but no significant increase in global emotional disturbance, compared to children. Blair et al. [3] found increased emotional disturbance with increased age, which did not appear to be the result of changing disease severity.

There are few papers dealing with CF in older adults because it is only recently that survival into adulthood has become common. Research is needed into the psychological and psychosocial impact of CF upon older adults and the research involving adolescents and younger adults needs to be repeated.

6. The effect of coping style

People with CF can be divided into copers and non-copers. It is hard to define the cut-off between coping

and non-coping, and similarly between different coping strategies. This leads to problems in comparing different studies that have used different methods of classifying coping.

Coping strategies can be subdivided into two groups—attention responders and avoidance responders. Avoidance may also be referred to as denial or minimisation. Most research concludes that attention responders may have a poorer psychological outcome than avoidance responders [26]. Strauss and Wellisch [27] reported ‘prominent use of denial’ and ‘minimisation’ in cystic fibrosis.

Staab et al. [12] found, using multiple regression analyses, that ways of coping contributed significantly to the overall variance in quality of life. Coping by using (positive) social comparison correlated with a higher quality of life while depressive coping was negatively correlated with quality of life. There was a weak correlation between cognitive avoidance and a worse quality of life. This study also found that the ways in which parents cope is an extremely important factor in determining the quality of life of the parents of children with cystic fibrosis (and possibly parents of CF adults).

Those who cope with avoidance generally have a worse compliance. Abbott et al. [28] found that patients adherent to treatment plans used the coping strategies of optimistic acceptance and hopefulness to a greater extent than those who were non-adherent, while the latter group of patients reported using avoidance strategies to a greater extent.

There needs to be more research in the area of coping styles. The question of which group has a better quality of life is unanswered. There is little, if any, information comparing the longer-term outlook (in terms of CF severity and life expectancy) of the two coping styles. Helping the patient achieve a beneficial coping strategy may be just as important as other forms of treatment.

7. Daily life and platonic relationships (fitting into society)

Many people with cystic fibrosis have problems with interpersonal relationships (both platonic and non-platonic) resulting in isolation and social maladjustment [29]. If relationships are not formed in teenage years they may never be formed [19].

Many people with CF experience problems with independence, such as fear of moving away from home [20]. A higher proportion of adults with CF than average remain living in the parental home in adulthood, especially men [8].

CF patients can suffer psychological problems of embarrassment of their cough, sputum and offensive stools when treatment is not working [30].

There is the anxiety that if their diagnosis is revealed, then regardless of their current health they may be refused employment [19]. Unemployment, which is not uncommon in CF, may cause psychological problems. The increasing survival of patients into adulthood, the advent of new and often expensive treatments and the lack of appreciation of CF as a disability mean that economic strain is becoming an increasing problem for CF patients although this differs greatly between countries.

8. Sexuality and fertility

Problems in this area include body image, intimacy, reduced fertility in both sexes, the hereditary nature of the disease and the fear of dying before any children that they have grow up [20,31,32]. Cystic fibrosis (per se) does not produce sexual difficulty until it becomes severe [33]. However, sexual problems have been found to be frequent amongst people with CF. There is an increased level of sexual dissatisfaction primarily among unmarried people with CF [4].

With the increasing survival of CF patients into adulthood with reasonable health, making pregnancy less dangerous, having children is becoming increasingly relevant. This is creating new possible psychological problems for people with CF requiring education and counselling [22]. Artificial insemination has been found to improve fertility. Bilateral absence of the vasa deferentia occurs in most men with cystic fibrosis, causing infertility. Although, donor sperm may circumvent male infertility, the technique of intracytoplasmic sperm injection (ICSI), which could be combined with pre-implantation genetic diagnosis if the partner is a carrier of a mutated CF gene, can now allow CF men to father children of their own [34]. However, this has the issues of any new technique, especially in the field of fertility studies, such as any adverse effects on health of progeny of ICSI in which the natural physiological selection of the fittest sperm does not occur [35]. Also there are those concerns that anyone with CF will have when considering having children, such as concerns of passing the disease responsible genes to offspring and the high possibility of being ill or dying during their progeny's childhood.

9. The family

Although studies in this area centre on children, as adolescents and even, at times, adults with CF remain within the family unit it is germane to discuss this here. Mothers have often been found to be overly involved and protective of the child with CF, while the fathers help less often leading to the father's withdrawal [36]. Depression is common among mothers (approx. 30%), but is far less common among the fathers [3,37]. In

1981, Bywater [37] noted that the majority of mothers said they would welcome help dealing with these psychological problems but had not been offered it. Siblings (without CF) of children with CF have been observed to be angry and resentful of the (comparative) lack of parental attention and showed more school problems [36]. This can lead to a family unit breakdown in areas of communication, social isolation, decreased satisfaction and poor adjustment [36].

It is important to note that family coping affects the health of the child with CF and possibly the health of people with CF when they are older—better family coping leads to better health of the child [36]. Such family coping involves maintaining an integrated, coherent family with good self-esteem. A decline in the health of the child with CF has been found to lead to increased family stress [36].

10. Disease understanding and treatment

Inadequate knowledge and incorrect beliefs about CF may be associated with psychological distress. There is a disparity between perceived and actual health and vulnerability. This indicates some uncertainty about the effects of the disease and/or lack of understanding about medical therapy [38].

In adults there is a common misconception that they are healthier than they are [14], though this does not mean that they may not overestimate some of the effects of the disease and wrongly attribute problems to being caused by their CF. This leads to normal problems/crises of life being wrongly attributed to having CF. It is important to help patients express secret worries and anxieties with counselling [39]. Adolescents and their parents also often show a tendency for overestimation, rather than under-estimation of the effects of the disease [39].

There are also possible psychological problems resulting from misunderstanding disease complications and treatment, especially rarer complications [40]. Some people with CF have problems dispelling irrational fears such as the expectation of a sudden death [41]. Strange physical complaints (e.g. somatic complaints not related to physical parameters) may be of emotional meaning [16].

Transplants and transplant waiting lists can lead to their own psychological problems. Patients suffer psychological problems owing to 'waiting' on a transplant list or being unsuitable for a transplant. One stress which may be suffered by transplantees is that someone else had to die for them to live [42]. A major reason for psychiatric referral of CF patients is assessment for suitability for transplant from a psychiatric perspective, for example in terms of coping, possible psychiatric problems and compliance. The new technique of lobe transplants from live donors may cause new psycholog-

ical problems, both for the CF patient and potential donors. For example, both recipient and donors may feel undue guilt and responsibility regarding any significant complications that might arise to the others involved.

Large improvements in quality of life have been shown following heart–lung transplantation in patients with CF [43]. The sensitivity of QOL scales to more minor CF treatments is uncertain. It is important that as new treatments such as gene therapy become available that before-and-after quality of life research is conducted, helping to assess the benefits of these treatments.

11. Non-compliance

There is a high-degree of non-compliance, with over half of people with CF showing significant non-compliance in at least one aspect of management [44]. Reasons for non-compliance can be divided into general factors that are not specific to particular treatments and factors that are treatment-specific.

Non-compliance may be accidental or intentional. Koocher et al. [45] divided reasons for non-compliance into inadequate knowledge, psychosocial resistance [struggles for control, peer group concerns, avoidance (coping style), despair] or educated non-adherence (choice based on a full understanding of reasons and results of following/not following the treatment). However, the most common cause of non-compliance may be forgetfulness [14]. It is important to acknowledge that incomplete adherence is normal and not to take a judgmental approach when dealing with the patient [46].

Many patients underestimate the severity of their CF especially as it deteriorates and they overestimate the quality of their self-care [13]. Such misvaluation may lead to the belief that compliance is not as important as it is, leading to increased non-compliance. Poor knowledge about treatment may also result in unintentional poor compliance with treatment regimes [40]. Treatment details are often misinterpreted; clear simple protocols are needed [47]. Compliance may be improved by both a better patient understanding of CF and the value of compliance.

There is a problem with chronic diseases that patients learn a lot about their disease and may disagree with their doctors about their treatment. The ensuing struggle for control may lead to non-compliance [25]. Non-compliance in general may result when doctors expect patients to submit to their authority and do not appear responsive to patient's beliefs and concerns, i.e. a bad doctor–patient relationship can lead to non-compliance [25].

Non-compliance is often associated with a coping mechanism of avoidance. Patients who have more outpatient visits have been found to be more compliant [14].

Abbott et al. [48] have found that the greater the level of worry regarding the disease, the more likely they were to adhere to their physiotherapy, pancreatic enzymes and vitamins but not exercise (which seems to be perceived differently). They also found that (reported) adherence to physiotherapy, exercise, pancreatic enzymes and vitamins is not influenced by patient's perceptions of their past, current and future disease severity or perceived susceptibility to recurrent infections, although other studies disagree with this. Perception of having little self-control over the disease has also been linked to greater (reported) adherence (except with exercise) [48]. If the level of perceived severity becomes too great, denial may begin [48].

Non-compliance may result when the patients find that the demands of CF treatments have to be balanced with those of other aspects of their lives, such as work, relationships and family [14]. This is again intentional non-compliance but of a more conscious, reasoned nature. Those feeling in control may deliberately decide not to comply because they perceive themselves to be well and in control, balancing treatment with living a normal life [48]. Non-compliance is more likely when treatments are incompatible with the person's goal of normality [25].

Compliance is generally worse in adulthood than childhood and may be worse in women than men. The time and commitment demands become more difficult in adolescence and adult life which may in part help to explain why compliance tends to decrease with age [14]. Another reason is that children are forced to comply with treatment by their parents. Problems of compliance are particularly likely during adolescence (due to the 'rebellious' nature of adolescents) [24,49]. Adolescents resent being helped by parents with physiotherapy, so Forced Expiration Technique in conjunction with postural drainage is a useful method of physiotherapy. Compliance is not influenced by demographic details, nor intelligence [47].

In many cases reasons for non-compliance are treatment-specific and compliance may be improved by more tailoring of treatment to a specific patient's life-style [14,44]. Compliance with vitamin treatment and physiotherapy is worse than with enzyme and exercise therapies [44]. Example reasons for non-compliance with particular treatments include [14]: embarrassment with taking pancreatic enzymes; exercise was a preferred alternative to physiotherapy; nebulised antibiotics and inhaled steroids were found to taste awful. Physiotherapy, dietary supplements and nebulised therapies require the most planning and are most interfering with life [14]. Physiotherapy was viewed as a large demand on time and commitment, repetitive and onerous, and coupled to the perception in some patients that it makes no difference, results in non-compliance. Exercise allows

social integration and compliance is therefore greater [44,47].

Compliance is extremely high with self-administered intravenous antibiotics taken at home which improves independence and causes less disruption and is therefore preferred to inpatient infusion [50].

Compliance is better when immediate and obvious benefits are experienced [25,44,47]. Self monitoring of respiratory function would be beneficial as it would indicate the benefits of medications, convincing patients that actions make a difference [14].

Lask [46] lists several important approaches to managing compliance: empathy (a warm, non-judgemental approach, acknowledging that incomplete compliance is normal); enthusiasm (communicating confidence and hope); exploration (of the individual's reasons for non-compliance and problems with treatments allowing a personalised treatment plan); education; and expression of emotion. It is important to allow and aid the person to express feelings such as sadness, anger, envy, fear, frustration, resentment and depression. Management is best when it is a therapeutic alliance between the patient and medical staff, the latter being friendly, empathic, supportive, non-critical and understanding [51].

Small group interactive sessions may be beneficial in cystic fibrosis, including managing poor compliance, providing peer support and the knowledge that others have had similar problems [14,52].

12. Conclusion

Cystic fibrosis is an uncommon but deadly disease. New treatments for cystic fibrosis are emerging including lobe transplants from live donors, gene therapy and new fertility treatments to allow males with CF to become biological fathers. Although life expectancy has increased, it is very important that we cater not just for quantity but also quality of life. In addition, prognosis is probably closely related to compliance which itself is greatly affected by psychosocial factors.

The psychological and psychosocial functioning of this group of patients appears to be relatively normal but there are aspects in which they have significant distress and disability. However, there is relatively little research into the psychological well being of adults with cystic fibrosis and with increasing numbers of patients surviving into adulthood, more research is needed. Furthermore, older studies are becoming increasingly inapplicable as treatment and prognosis changes.

Research is needed to clarify whether mental health problems are a significant factor in patients with cystic fibrosis and if so whether they need treatment in their own right and whether such treatment affects the prognosis of the underlying medical condition. If poor mental health is a significant factor then research is needed into what are the predictors of significant mental health

problems and what preventative measures can be taken. For example should routine psychological assessment be carried out at regular intervals in cystic fibrosis and should specialised psychiatric input be available to CF units?

References

- [1] Anderson DL, Flume PA, Hardy KK. Psychological functioning of adults with cystic fibrosis. *Chest* 2001;119:1079–84.
- [2] Pearson DA, Pumariega AJ, Seilheimer DK. The development of psychiatric symptomatology in patients with cystic fibrosis. *J Am Acad Child Adolesc Psychiatry* 1991;30(2):290–7.
- [3] Blair C, Cull A, Freeman CP. Psychological functioning of young adults with cystic fibrosis and their families. *Thorax* 1994;49:798–802.
- [4] Shepherd SL, Hovell MF, Harwood IR, et al. A comparative study of the psychosocial assets of adults with cystic fibrosis and their healthy peers. *Chest* 1990;97(6):1310–6.
- [5] de Jong W, Kaptein AA, van der Schans CP, et al. Quality of life in patients with cystic fibrosis. *Pediatr Pulmonol* 1997;23:95–100.
- [6] Congleton J, Hodson ME, Duncan-Skingle F. Quality of life in adults with cystic fibrosis. *Thorax* 1996;51(9):936–40.
- [7] Britto MT, Kotagal UR, Hornung RW, Atherton HD, Tsevat J, Wilmott RW. Impact of recent pulmonary exacerbations on quality of life in patients with cystic fibrosis. *Chest* 2002;121:64–72.
- [8] Walters S, Britton J, Hodson ME. Demographic and social characteristics of adults with cystic fibrosis in the United Kingdom. *B M J* 1993;306(6877):549–52.
- [9] Orenstein DM, Nixon PA, Ross EA, Kaplan RM. The quality of well-being in cystic fibrosis. *Chest* 1989;95(2):344–7.
- [10] Orenstein DM, Pattishall EN, Nixon PA, Ross EA, Kaplan RM. Quality of well-being before and after antibiotic treatment of pulmonary exacerbation in patients with cystic fibrosis. *Chest* 1990;98(5):1081–4.
- [11] Congleton J, Hodson ME, Duncan-Skingle F. Do Nottingham Health Profile scores change over time in cystic fibrosis? *Respir Med* 1998;92:268–72.
- [12] Staab D, Wenninger K, Gebert N, et al. Quality of life in patients with cystic fibrosis and their parents: what is important besides disease severity. *Thorax* 1998;53:727–31.
- [13] Abbot J, Dodd M, Webb AK. Different perceptions of disease severity and self care between patients with cystic fibrosis, their close companions and physician. *Thorax* 1995;50(7):794–6.
- [14] Conway SP, Pond MN, Hamnett T, Watson A. Compliance with treatment in adult patients with CF. *Thorax* 1996;51(1):29–33.
- [15] Gee L, Abbott J, Conway SP, Etherington C, Webb AK. Development of a disease specific health related quality of life measure for adults and adolescents with cystic fibrosis. *Thorax* 2000;55:946–54.
- [16] Simmons RJ, Corey M, Cowen L, Keenan N, Robertson J, Levinson H. Emotional adjustment of early adolescents with cystic fibrosis. *Psychosom Med* 1985;47(2):111–22.
- [17] Willis E, Miller R, Wyn J. Gendered embodiment and survival for young people with cystic fibrosis. *Soc Sci Med* 2001;53:1163–74.
- [18] Nicholson B. My experience as an adult with cystic fibrosis. *J R Soc Med* 1993;86(Suppl 20):30–3.
- [19] Mearns MB. Special problems for the teenager with cystic fibrosis. *J R Soc Med* 1986;79(Suppl 12):51–4.

- [20] Kollberg H. Cystic fibrosis in adulthood. *Eur J Respir Dis Suppl* 1982;118:101–9.
- [21] Landon C, Rosenfeld RG. Short stature and pubertal delay in cystic fibrosis. *Pediatrician* 1987;14(4):253–60.
- [22] Johannesson M, Carlson M, Brucefors AB, Hjelte L. Cystic fibrosis through a female perspective: psychosocial issues and information concerning puberty and motherhood. *Patient Educ Couns* 1998;34(2):115–23.
- [23] Raymond NC, Chang PN, Crow SJ, et al. Eating disorders in patients with cystic fibrosis. *J Adolesc* 2000;23:359–63.
- [24] Landau LI. Cystic fibrosis: transition from paediatric to adult physician's care (editorial). *Thorax* 1995;50(10):1031–2.
- [25] Wright AL, Morgan WJ. On the creation of 'problem' patients. *Soc Sci Med* 1990;30(9):951–9.
- [26] Aspin AJ. Psychological consequences of cystic fibrosis in adults. *B J Hosp Med* 1991;45(6):368–71.
- [27] Strauss G, Wellisch D. Psychosocial adaptation in older cystic fibrosis patients. *J Chronic Dis* 1981;34:141–6.
- [28] Abbott J, Dodd M, Gee L, Webb K. Ways of coping with cystic fibrosis: implications for treatment adherence. *Disabil Rehabil* 2001;23(8):315–24.
- [29] Sinenema G, Bonarius JC, Stoop JW, van der Laag J. Adolescents with cystic fibrosis in the Netherlands. *Acta Paediatr Scand* 1983;72(3):427–32.
- [30] Hodson ME. Psychological and social aspects of cystic fibrosis. *Practitioner* 1980;224(1341):301–3.
- [31] Elliott M. Cystic fibrosis. *Practitioner* 1989;233(1463):253–6.
- [32] Sinnema G, Van der Laag H, Stoop JW. Psychological development as related to puberty, body height and severity of illness in adolescents with cystic fibrosis. *Isr J Med Sci* 1991;27(4):186–91.
- [33] Levine SB, Stern RC. Sexual function in cystic fibrosis. Relationship to overall health status and pulmonary disease severity in 30 married patients. *Chest* 1982;81(4):422–8.
- [34] McCallum TJ, Milunsky JM, Cuningham DL, Harris DH, Maher TA, Oates RD. Fertility in men with cystic fibrosis. An update on current surgical practices and outcomes. *Chest* 2000;118:1059–62.
- [35] Kim ED, Bischoff FZ, Lipshultz LI, Lamb DJ. Genetic concerns for the subfertile male in the era of ICSI. *Prenat Diagn* 1998;18:1349–65.
- [36] Patterson JM, McCubbin HI, Warwick WJ. The impact of family functioning on health changes in children with cystic fibrosis. *Soc Sci Med* 1990;31(2):159–64.
- [37] Bywater EM. Adolescents with cystic fibrosis: psychosocial adjustment. *Arch Dis Child* 1981;56(7):538–43.
- [38] Brown C, Rowley S, Helm P. Symptoms, health and illness behaviour in cystic fibrosis. *Soc Sci Med* 1994;39(3):375–9.
- [39] Jedlicka-Köhler I, Götz M. Interventional assessment of physical and mental health in children and adolescents with cystic fibrosis. *Scand J Gastroenterol Suppl* 1988;143:34–7.
- [40] Conway SP, Pond MN, Watson A, Hamnett T. Knowledge of adult patients with cystic fibrosis about their illness. *Thorax* 1996;51(1):34–8.
- [41] Dushenko TW. Cystic fibrosis: a medical overview and critique of the psychological literature. *Soc Sci Med (E)* 1981;15(1):43–56.
- [42] Oakman P. My heart–lung transplant. *J R Soc Med* 1993;86(Suppl 20):34–5.
- [43] Caine N, Sharples LD, Smyth R, et al. Survival and quality of life of cystic fibrosis patients before and after heart–lung transplantation. *Transplant Proc* 1991;23:1203–4.
- [44] Abbot J, Dodd M, Bilton D, Webb AK. Treatment compliance in adults with cystic fibrosis. *Thorax* 1994;49:115–20.
- [45] Koocher GP, McGrath M, Gudas LJ. Typologies of non-adherence in cystic fibrosis. *J Dev Behav Pediatr* 1990;11(6):353–8.
- [46] Lask B. Non-adherence to treatment in cystic fibrosis. *J R Soc Med* 1994;87:25–7. Suppl 21.
- [47] Webb AK. Communicating with young adults with cystic fibrosis. *Postgrad Med J* 1995;71(837):385–9.
- [48] Abbott J, Dodd M, Webb AK. Health perceptions and treatment adherence in adults with cystic fibrosis. *Thorax* 1996;51:1233–8.
- [49] Hodson ME. Cystic fibrosis in adolescents and adults. *Practitioner* 1983;227(1385):1723–31.
- [50] Strandvik B, Hjelte L, Malmberg AS, Widen B. Home intravenous antibiotic treatment of patients with cystic fibrosis. *Acta Paediatr* 1992;81(4):340–4.
- [51] Lask B. Understanding and managing poor adherence in cystic fibrosis. *Pediatr Pulmonol* 1997;Suppl 16:260–1.
- [52] Strauss GH, Pedersen S, Dudovitz D. Psychological support for adults with cystic fibrosis. *Am J Dis Child* 1979;133(3):301–5.